Congenital Cystic Malformation of the Bile Ducts:
Report of a Case and Review of Related Literature

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A 20-year-old woman had a cyst of the proximal part of the common bile duct and a cyst of the left hepatic duct; these lesions were diagnosed preoperatively by intravenous cholangiography and successfully operated upon. At the time of writing, she has been followed up for one year.

Congenital defects in the biliary system are rare and, in a review of the literature, only two cases were found similar to this one. It is generally accepted that these lesions are congenital, but the exact pathogenesis is unknown.

Alonso-Lej, Rever and Pessagno[2] reviewed the literature in 1959 and found 403 authentic congenital cysts of the hepatic ducts. The most common congenital defect is a single choledochal cyst of the lower end of the common bile duct. Pain, jaundice and tumour are the main symptoms.

Until the advent of intravenous cholangiography, these lesions were seldom recognized preoperatively. Means of operative repair as well as complications and prognosis are reviewed.

On a constaté chez une femme de 20 ans un kyste de la portion proximale du cholédoque et un kyste du canal hépatique gauche. Ces lésions ont été diagnostiquées avant l'opération, grâce à une cholangiographie intraveineuse, et ont pu être opérées avec succès. Au moment où l'auteur a écrit le présent article, la patiente a été suivie pendant un an.

Les défauts des voies biliaires d'origine congénitale sont rares, à tel point qu'une revue complète de la littérature n'a permis de déceler que deux cas similaires au présent cas. On admet généralement que ces lésions sont congénitales, bien que leur pathogénie demeure obscure.

Alonso-Lej, Rever et Pessagno[2] ont passé en revue la littérature en 1959 et ont découvert 403 cas authentiques de kystes congénitaux des canaux hépatiques. Le défaut congénital le plus courant est un kyste simple de la portion inférieure du cholédoque. Les principaux symptômes sont douleur, ictère et gonflement.

Jusqu'à l'avènement de la cholangiographie intraveineuse, il était rare qu'on découvra ces lésions avant l'intervention. Les modes opératoires, les complications et le pronostic sont passés en revue.

CONGENITAL cysts of the bile ducts have long been known. Vater and Ezler[1] described the first case of a congenital cyst of the common bile duct in 1723. Alonso-Lej, Rever and Pessagno[2] gathered 403 authentic cases of choledochal cysts from the world literature up to 1959. The present communication describes a 20-year-old woman with a cyst of the proximal part of the common bile duct and a cyst of the left hepatic duct, which were diagnosed preoperatively by intravenous cholangiography and successfully operated upon one year ago.

CASE REPORT

Miss J.M., aged 20, was admitted to the Toronto Western Hospital, August 7, 1964, with the chief complaint of right upper quadrant pain accompanied by nausea and vomiting. The pain was constant, not crampy, coming on slowly and building to a maximum over two to three hours.

Examination on admission revealed a thin, pale girl weighing 87 lb., 5'2" in height, in some distress. Ears, nose and throat were clear. There was no jaundice. Chest and breasts were normal. Blood pressure was 118/75 mm. Hg. The pulse 78 per minute and regular. The heart was normal in size and there were no murmurs. The liver edge was just palpable at the right costal margin; no other organs or masses were felt. There was slight tenderness in the right upper quadrant, and the scar of a previous appendectomy in the right lower quadrant. Rectal examination was negative. The patient was alert, co-operative, and of average intelligence, although quite apprehensive. Cranial nerves were intact and reflexes all present and equal. Sensation was intact.

The pain had come on 48 hours before admission. The patient had been given 100 mg. of meperidine hydrochloride by her family doctor the night before admission, and the pain had settled down to a dull ache. Enquiring into her past health, it was learned that in infancy she had been jaundiced for a short period during her first six months of life but recovered in about 10 days with no investigation or hospitalization. Her mother said she was always "delicate", underweight, nervous, and easily upset by rich or fatty foods which caused vomiting.

In December 1954 she had an episode of jaundice associated with abdominal pain and vomiting, with dark urine and pale stools, and was treated at home and recovered in about three weeks. On March 15, 1955, at the age of 11 she was admitted to The Hospital for Sick Children, Toronto, because of abdominal pain and vomiting. She was thought to have acute appendicitis and an appendectomy was performed. The day after operation, she was seen to be jaundiced. On this admission an intravenous cholangiogram was performed and this revealed a peculiar concentration of dye in what appeared to be a choledochal cyst. Nothing further was done at this time and she recovered uneventfully.

She remained well until August 1961, when she again had upper abdominal pain and vomiting and was admitted to a private hospital in Toronto. No details of that admission are available but the patient says she was not jaundiced. She recovered and remained well until the present admission.

Laboratory examination on this admission revealed a hemoglobin of 12.7 g. %, white blood count of 5600/c.mm. with normal differential blood count, and

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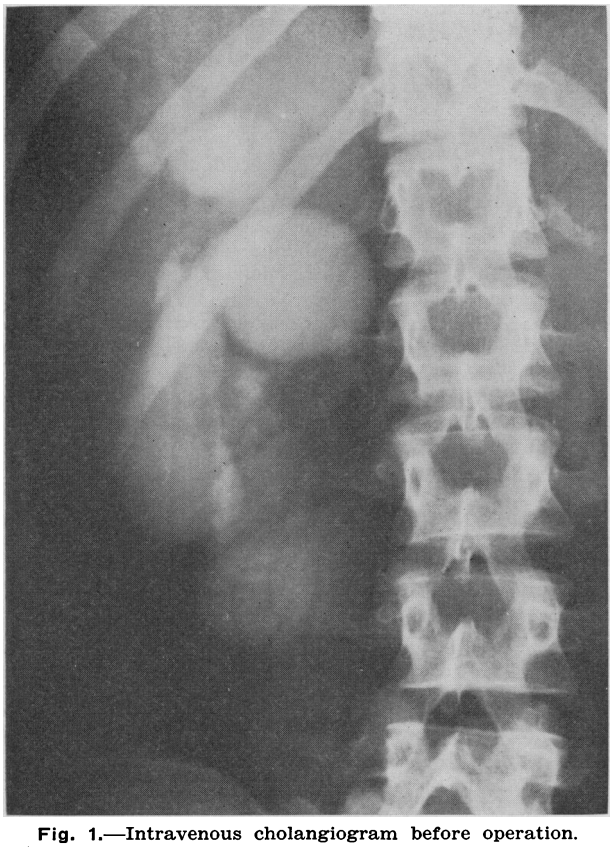

Fig. 1.—Intravenous cholangiogram before operation.

normal blood smear. Sedimentation rate was 14 mm./hr. The urine had a specific gravity of 1.018, was negative for bile, but showed a urobilin 1 plus. The bromsulphalein test (BSP) showed 5% retention at 45 minutes. The direct bilirubin was 0.08 mg. % with a total of 0.76 mg. %. Serum glutamic oxaloacetic transaminase (SGOT) was 4 units, cephalin-cholesterol flocculation 2 plus, alkaline phosphatase 7.8 King-Armstrong units, serum amylase 142 units, serum sodium 136 mEq./l., serum potassium 4.1 mEq./l., serum chlorides 90 mEq./l., CO_2 combining power 26.1%, and serum proteins 6.5 g. % with an albumin of 4.38 g. % and normal globulins on electrophoresis. Her body temperature was normal.

On iopanoic acid (Telepaque) examination, the gallbladder, which was faintly outlined, was of normal size, shape and position. A rounded 4 cm. dye-containing density was recognized adjacent to the upper part of the gallbladder. To give further detail, an intravenous cholangiogram was done. (We did not have details of the Sick Children's Hospital admission at this time, or until after subsequent operation.) On intravenous cholangiogram the gallbladder filled well and appeared normal. However, adjacent to the upper end of the gallbladder and in the expected position of the superior part of the common bile duct, a round dye-filled 5 cm. density was recognized. From the lower part of the density, dye passed inferiorly into the middle and the lower one-third of the common bile duct, which appeared normal in calibre (Fig. 1). The distal 1 cm. of the common bile duct was not visualized and there was a slow flow of contrast medium into the duodenal loop, which did not appear remarkable. Above the common bile duct and the rounded density already described, there were at least two other rounded densities. There was some obstruction to free flow of contrast medium out of the biliary system and, during this examination, the patient had severe pain and vomiting. In the radiologist's opinion, there was dilatation of several segments of the biliary tree including the proximal common bile duct and the distal parts of the hepatic ducts in the region of the porta.

The patient underwent operation on August 14. The gallbladder was normal. There was a large cystic swelling of the upper portion of the common bile duct and, between this and the second cystic swelling of the left hepatic duct, there was a stricture which barely admitted a No. 12 dilator. After an operative cholangiogram was done, the cystic artery was ligated and divided for exposure. The narrow constriction between the two cystic swellings was cut longitudinally. A No. 18 T-tube was introduced and a portion of the lower choledochocyst was brought up into the stricture for closure in an oblique fashion. The duodenum proximal to the lower cyst was opened and a choledochoduodenostomy was carried out. A T-tube was left in place. The patient withstood the procedure well, and made an uneventful recovery.

A T-tube cholangiogram on August 24 (Fig. 2) showed a dilated area near the junction of the right and the left branches of the bile duct. Distal to this, there was a narrowed area of the bile duct and then another dilated area. From this second dilated area, the cystic duct and the gallbladder filled and the second dilated area was that anastomosed to the first part of the duodenum. No calculi were seen and the dye drained well into the duodenum. The T-tube was removed on August 31. The patient did well. Liver function tests remained normal throughout her stay in hospital.

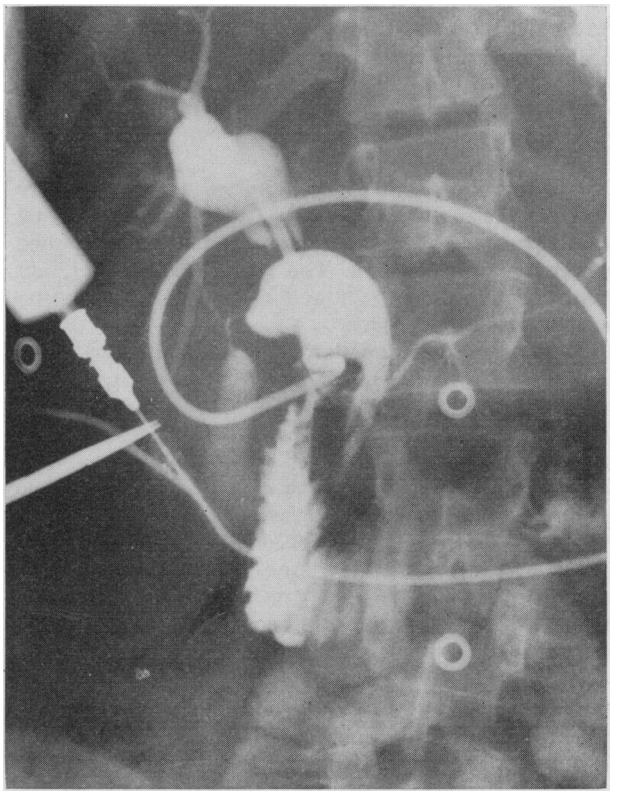

Fig. 2.—T-tube cholangiogram postoperatively.

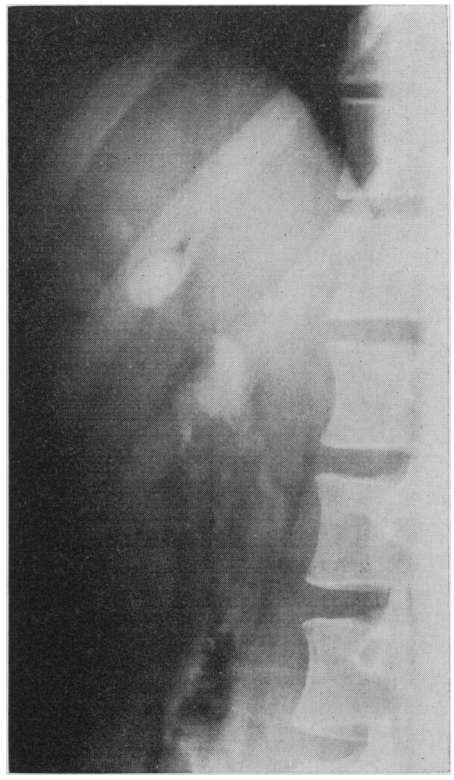

Fig. 3.—Intravenous cholangiogram one year after operation.

The patient was seen again in October, 1964. She was feeling well and had had no recurrent pain, nausea, vomiting or jaundice. Physical examination was completely normal. She was seen again in April 1965 and again was well. A BSP done at this time showed 4.8% retention at 45 minutes, and a repeat intravenous iodipamide (Cholographin) examination was carried out on April 23, 1965 (Fig. 3). There was prompt function. The lower end of the common bile duct was normal, and there was a free flow of the medium into the small bowel through the anastomosis. There was far less evidence of dilatation of the cystic structures than at previous examination.

MALFORMATIONS OF THE BILIARY TREE

Classification.—Those in categories I-III (Table I) occur in infancy, usually within two or three weeks of birth and are associated with jaundice. Some are amenable to surgical repair. The actual incidence or occurrence of the lesions in category IV is uncertain. Those in category V may be congenital but are most often due to previous surgery or infection. Cattell[3] described 123 patients operated upon for stricture of the biliary ducts before 1945 and 30 operated upon after 1945. The usual cause of stricture in this group was previous surgery. The causes of the injury were operative in 99, cholangitis in 11, fibrosis of the ampulla of Vater in nine, adhesions in two, trauma in one, and one unknown. The last category, VI, was added by Arthur and Stewart[4] to include their patient as well as that of McWhorter,[5] and would also cover our own patient.

Etiology.—The conditions listed in Table I are presumed to be congenital in origin, when the following factors have been ruled out: previous disease of the biliary tract, such as cholangitis or cholelithiasis; if, at operation, no stones are found that could have caused ductal injury; if no previous biliary tract operation has been performed; if there is no evidence of adjacent organ disease, and no history of abdominal trauma or other abdominal surgery.

TABLE I.—CLASSIFICATION OF MALFORMATIONS OF THE BILIARY TREE ACCORDING TO HOLMES,[23] MODIFIED BY ARTHUR AND STEWART[4]

I.	Atresia: (a) Those in which there are no extrahepatic ducts. (b) Atresia of the hepatic ducts. (c) Atresia of the common bile duct.
II.	Those in which the gallbladder is represented by a moderate-sized cyst that does not connect with the common bile duct, and in which there may or may not be any common or hepatic ducts.
III.	Those in which the gallbladder connects directly with the duodenum, but in which there are no extrahepatic ducts, that is, no ducts connect the liver and the gallbladder, or the liver and the intestine.
IV.	Stenosis of the common duct with inspissated bile causing complete obstruction.
V.	Those in which there is a narrowing of the common duct causing partial obstruction.
VI.	Multiple biliary cysts.

Waller[6] quotes Heiliger who found multiple biliary cysts in a stillborn fetus. This observation tends to confirm the general opinion that these cysts are congenital. Thomson[7] collected 49 cases of choledochal cysts in 1891 and added one of his own. He noted that these cysts had a familial incidence.

It is interesting to speculate how these defects may develop. According to Arey,[8] the main ducts and the gallbladder are subject to duplication as the result of early splitting, subdivision or sacculation, and a congenitally narrowed or solid condition of the gallbladder or of the chief ducts represents persistence of embryonic occlusion. Hence atresia can be readily explained. Cysts of hepatic ducts could result from sacculation during the embryonic stage. According to Ylppö,[9] in the process of development the bile ducts pass through a solid state and the lumen of the ducts becomes obliterated by epithelial concrescences or proliferation. If development is normal, they go on to form solid cords which become vacuolated. The vacuoles coalesce and the lumen is re-established.

Tsardakas and Robnett[10] and Alonso-Lej, Rever and Pessagno[2] believe that the condition is congenital. They propose that weakness of a specific portion of the common bile duct is due to hyperproliferation and hypervacuolization of that area during embryonic development. When ductal pressure increases following obstruction, this area dilates. They class cystic dilatations of the bile ducts as follows: congenital cystic dilatation, congenital diverticulum of the common bile duct, and congenital choledochocele. The first type is the most common.

Incidence.—The true incidence of congenital malformations of the bile ducts is unknown.

Sweet[11] in his paper "Congenital Malformation of the Bile Ducts", 1933, described three cases in one family. All these were due to ductal atresia. Among 45,000 cases admitted to the St. Louis Children's Hospital, Cole[12] found only six children with congenital malformation of the bile ducts. Judd and Green[13] reported that, in 17,381 operations on the biliary tract at the Mayo Clinic, a true cyst of the common bile duct was found only once. Ladd,[14] at the Children's Hospital in Boston, had 45 patients with congenital deformities of the bile ducts; of these 27 had inoperable conditions, and the usual duration of life was four to five months. Twenty were explored and found amenable to surgical treatment. According to Smith,[15] among 757,000 admissions to the Presbyterian Hospital in New York City, only two cases of cysts of the common bile duct were found. Among Caucasians, the cyst is four times commoner in the female than the male. One-third of reported cases involve Japanese, in whom the sex incidence is equal. Arthur and Stewart[4] state that three-quarters of the cases are under 25 years of age but individuals of any age may be affected. Daniel[16] believes that the process is usually discovered early; 85% of the patients he reviewed were hospitalized before the age of 40, and 45% before the age of 10.

Diagnosis.—The diagnosis of congenital cysts of the bile ducts was difficult preoperatively until recently when cholecystography was introduced. Moseley[17] in 1957 described the first such case demonstrated by oral cholecystography; the next was recorded by Paulino[18] in 1957. Hankamp[19] in 1959 described four cases diagnosed by cholecystography. Ferris and YaDeau[20] at the Mayo Clinic, in 1964 reported four cases of choledochal cysts, three of which were visualized radiographically before operation. Tsardakas and Robnett[10] believe that the diagnosis is often difficult and suggest that the new intravenous dyes offer the best chance of diagnosing this rare condition because, following oral cholecystography, the area of cystic dilatation often does not concentrate the opaque medium.

Signs and Symptoms.—The signs and symptoms pointing to the diagnosis of congenital malformations of the biliary tree are many and varied, but most authors have noted the presence of pain, jaundice, mass in the right upper quadrant, nausea and vomiting. Cole[12] found that the symptoms of congenital malformation of the bile ducts and of cystic dilatation of the bile ducts were variable. The condition is usually found in a person about 18 years of age who has symptoms referable to the biliary tree, e.g. nausea, pain and jaundice. Occasionally a tumour can be palpated in the right upper quadrant. Zinninger and Cash,[21] reviewing the literature, found 109 cases of cystic dilatation of the extrahepatic ducts. Eighty-three of these were available for study; the common symptoms were jaundice and abdominal pain; some cases had tumour. The symptoms were usually present in infancy and childhood; they disappeared only to reappear later. Many had intermittent attacks over many years. The patients were predominantly female; in Zinninger's group, 68 of the 83 were females. In his experience, cystic dilatation of the hepatic ducts is a disease of children and young adults. The true diagnosis preoperatively, until the advent of cholecystography, was usually obscure. Shallow, Eger and Wagner[22] described 182 authenticated cases of congenital cystic dilatation of the common bile duct, in which the triad of symptoms: abdominal pain, mass in the right upper quadrant and jaundice, was prominent. He analyzed 75 of these cases: 77% had an abdominal mass, 70% jaundice and 59% abdominal pain.

Treatment and Prognosis.—Until recently the treatment and prognosis in congenital malformations of the biliary tree were disappointing. It was not until 1916 that Holmes[23] recommended that these patients should be operated upon, if at all possible, so that the diagnosis could be established and treatment, if any, could be carried out. Cole[12] described six patients with congenital malformation of the bile ducts; in one of these, operation was successful. Zinninger and Cash[21] in a review of the literature found 83 such cases. The preoperative diagnosis was correct in only three of these and the mortality in 54 patients operated upon was 65%. Shallow, Eger and Wagner[22] report a 51% operative mortality in their review. Tsardakas *et al.* reported on 52 patients with congenital cystic dilatation of the common bile duct who were operated upon with a mortality of 23%. Alonso-Lej, Rever and Pessagno,[2] in their study of congenital choledochal cysts in 1959, report an operative mortality of 15%.

Treatment of congenital malformations of the biliary tree was well described by Madding[24] in 1961. He discusses the following procedures:

1. External drainage or marsupialization. Arthur and Stewart[4] believe that this procedure should be abandoned. The risk of infection is very great.

2. Sphincterotomy alone, or with another procedure. This is seldom performed today.

3. Plastic repair or excision of the cystic dilatation with reconstruction of the common bile duct. If feasible, this is probably the best form of treatment. According to Arthur and Stewart,[4] the fate of the congenital cystic dilatations of the biliary system is uncertain after any bypass. They have concluded that the cysts never disappear completely unless excised, and that bypasses often fail because even an adequate anastomosis may become stenosed.

4. Bypass procedures joining the gallbladder to stomach, duodenum and jejunum. These have been highly popular when feasible.

5. Excision of the cystic dilatation has been advocated with choledochoduodenostomy or choledochojejunostomy (Roux-en-Y).

6. The final type of repair has been direct anastomosis of the cyst to the gastrointestinal tract: choledochocystoduodenostomy, choledochocystogastrostomy, and choledochocystojejunostomy; either Roux-en-Y type or a loop type with Braun anastomosis.

According to Gross,[25] 30% of patients who underwent choledochocystoduodenostomy had evidence of cholangitis or stenosis on follow-up. However, this procedure is a satisfactory alternative to excision of the cyst when the latter is not feasible. Choledochocystogastrostomy carries a low morbidity but a high mortality. In the experience of Alonso-Lej, Rever and Pessagno,[2] the mortality was 38%. According to the same author, cholecystojejunostomy with Roux-en-Y or a Braun-type anastomosis was introduced by the Japanese, but he states that it carries a very high mortality. In Alonso-Lej's series, five cases having had a bypass procedure had complete closure of the anastomosis. Ferris and YaDeau,[20] reporting from the Mayo Clinic, believe that a Roux-en-Y jejunal anastomosis is the procedure of choice because the risk of reflux (ascending) cholangitis is much less. Complete removal of multiple biliary cysts is usually not feasible unless they are all extrahepatic, but improvement of common bile duct drainage in such cases by an anastomatic procedure reduces morbidity from ascending cholangitis.

Complications.—The complications of biliary cysts are serious and include rupture with peritonitis; stone formation in the cyst; malignancy; ascending cholangitis before surgery and, if obstruction of the anastomosis occurs, subsequently after surgery; and biliary cirrhosis.

Discussion

Congenital malformations in the biliary system are rare. In a review of the literature we found only two cases comparable to our own. In 1939, McWhorter[5] at postmortem examination found multiple cysts involving the hepatic ducts, in a patient who had had a choledochal cyst removed 13 years before. Arthur and Stewart[4] in September, 1964, described multiple cystic dilatations of the intra- and extrahepatic radicals of the biliary system. They noted that most cysts are single choledochal cysts of the common bile duct and occur in the portion between the cystic duct and the first part of the duodenum. The patient they described had a cyst of the common bile duct as well as cysts of the right and the left hepatic ducts.

Choledochal cysts of the common bile duct have been recorded by many authors. Tsardakas and Robnett,[10] as recently as 1956, reported an operative mortality of 23% in attempts to repair congenital malformations of the extra-hepatic bile ducts.

Our patient had successful repair of the defects and one year later remains well. She has normal liver function and, by intravenous cholangiography, her anastomosis is widely patent and the cystic dilatations seen previously are considerably smaller; drainage of the biliary system into the duodenum is prompt. We feel that our patient falls in the group of congenital cystic dilatations of the common bile duct and hepatic ducts. She had no previous history of cholelithiasis or cholangitis, and no stones were found at operation which could have caused duct injury. She had never had previous biliary tract surgery and she had no evidence of any adjacent organ disease, no history of abdominal trauma or abdominal surgery in that area. She had no evidence of organic disease elsewhere in the body.

Acquired lesions of the biliary tract are much commoner than congenital malformations. The diagnosis and treatment of the acquired lesions is much more straightforward than that of the congenital lesions. Much has been written in the literature in the past two decades about the operative repair and handling of acquired stricture in the biliary tract, but there is still great confusion about the diagnosis and the handling of the congenital malformations.

Summary

A 20-year-old woman with congenital cysts of the upper part of the common bile duct and the left hepatic duct is described. The lesion was diagnosed preoperatively by intravenous cholangiography and successfully operated upon one year ago. At the time of writing, the patient remains well. Her liver function tests are normal and, by intravenous cholangiography, a widely patent choledochoduodenostomy is seen. A review of the literature including the etiology, pathogenesis, classification, incidence signs and symptoms, means of diagnosis and methods of treatment, as well as complications and prognosis, is presented.

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